Pneumatosis cystoides intestinalis in a patient with dermatomyositis

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J R Soc Med 1999;**92**:366–367

Pneumatosis cystoides intestinalis is a condition that tends to be diagnosed after lengthy investigations for non-specific gastrointestinal symptoms. An association with dermatomyositis has been reported but is rare.

CASE HISTORY

A woman aged 61 was referred with a 12-month history of burning upper abdominal pain, vomiting after meals and a 12 kg weight loss. Polymyositis had been diagnosed 18 months earlier and she was taking a maintenance dose of 12.5 mg prednisolone. She was obese and slightly tender in the epigastrium and right upper quadrant. Rebound tenderness and guarding were absent. No masses were palpable and bowel sounds were normal. Upper gastrointestinal endoscopy revealed a 5 cm hiatus hernia and atrophic gastritis (*Helicobacter pylori* negative) for which omeprazole 20 mg daily was prescribed. On ultrasound scanning there was no obvious hepatobiliary or pancreatic abnormality.

When reviewed after 4 weeks' treatment, she said that in the preceding week the pain had become worse, with abdominal distension. She was apyrexial and normotensive, heart rate 76 per minute. Her upper abdomen was again slightly tender, now with a resonant percussion note and high-pitched bowel sounds. Plain chest and abdominal radiographs showed free intraperitoneal gas, dilated small and large bowel, and pneumatosis cystoides intestinalis. She was managed non-operatively with intravenous fluids and antibiotics (cefotaxime 1 g and metronidazole 500 mg three times daily) and her pain resolved completely within 24 hours. Radiological contrast studies did not identify any site of gastrointestinal perforation. A computed tomogram revealed a loop of small bowel in the pelvis and a soft tissue mass in the right iliac fossa (Figure 1). It also showed calcification of subcutaneous tissues posterior

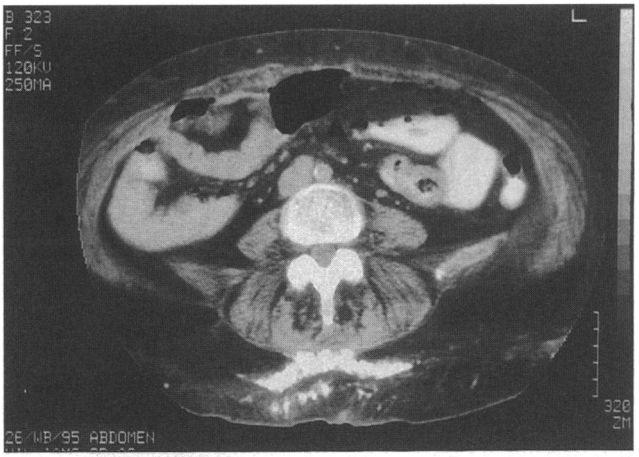

Figure 1 **Computed tomogram of abdomen.** Note a soft tissue mass in right iliac fossa together with calcification in skin overlying right side of the lumbar spine

to the lumbar spine, indicating skin involvement and signifying dermatomyositis rather than the previous diagnosis of polymyositis.

Continuing symptoms four months after initial presentation led to a precautionary laparotomy at which a band adhesion to an umbilical hernia repair scar was divided. For 70 cm proximal to this band, the ileal wall contained multiple cysts of variable size (Figure 2). Distal to the adhesion, for 70 cm the small bowel wall had a white fibrotic surface appearance but was devoid of cysts. There was a further area of florid cystic change in the distal 15 cm of ileum. No abnormality was identified in the colon or other intra-abdominal organs.

Division of the adhesion made little difference to the patient's symptoms and she continued to have episodes of abdominal pain and pneumoperitoneum. Subsequently an elemental diet gave temporary symptomatic relief. A further response was achieved with selective decontamination of the gut by a combination of oral colistin and neomycin; but one month after discontinuation of antibiotics the symptoms recurred and a second laparotomy was performed at which recurrent adhesions were identified in the region of the terminal ileum. The adhesions were divided and a 80 cm section of the affected distal ileum was resected with end-to-end anastomosis. The resected bowel showed typical histological features of pneumatosis cystoides intestinalis with a giant cell reaction around the gas filled cysts. When last seen, 13 months postoperatively, the patient was symptom-free.

COMMENT

The typical features of polymyositis are weakness and wasting of voluntary musculature, with pain and tenderness of proximal muscle groups. When skin changes

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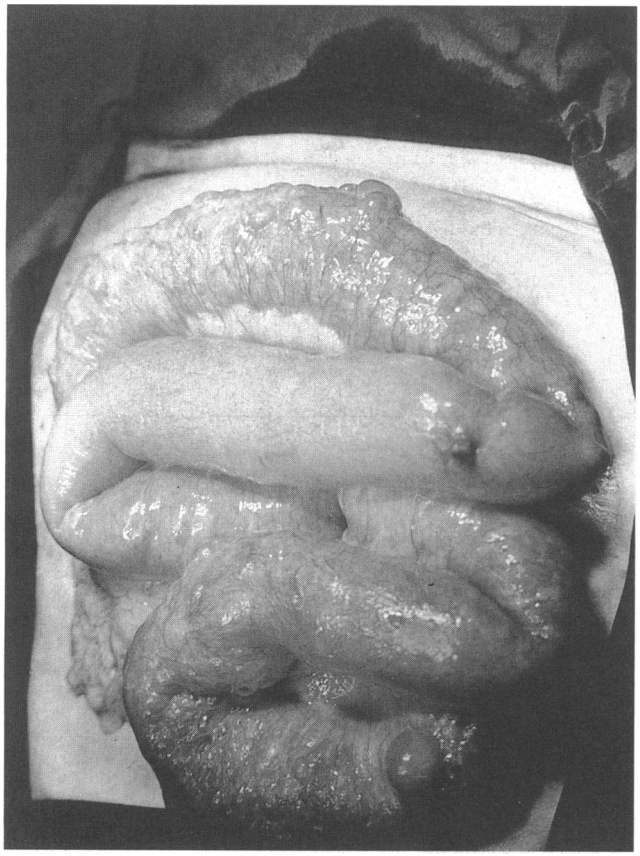

Figure 2 **Operative photograph showing cysts in wall of ileum**

are present, the condition is termed dermatomyositis. Both dermatomyositis and polymyositis can be associated with connective tissue disease or immunodeficiency, and in men over 45 years underlying malignancies are present in 50% of cases[1]. Pneumatosis cystoides intestinalis, likewise a rare condition of unknown aetiology, is secondary to another condition in 85% of patients[2].

Two theories have been proposed to account for the gas in the bowel wall in pneumatosis cystoides intestinalis. According to the mechanical hypothesis, gas is forced in via a breach in the mucosa such as might occur in peptic ulceration, or after traumatic instrumentation, or at a bowel anastomosis, or proximal to an obstructing lesion; or it might enter via the mesentery when, after alveolar rupture, it tracks though the mediastinum to the peritoneum[2]. The bacterial theory, that the gas arises from bacterial metabolism, is supported by the finding of high concentrations of hydrogen both within the cysts and in patients' breath[3]. Moreover, Yale *et al.* produced a similar condition in rats by injection of *Clostridium perfringens* either intraperitoneally or into the bowel wall[4].

There are only five previous reports of pneumatosis cystoides intestinalis in conjunction with dermatomyositis[5–9], and four of them[5–8] concerned children. The pathogenic link, if there is one, may be intravascular thrombosis, since in childhood dermatomyositis there is inflammation of the intramuscular arterioles with associated thrombosis and infarction (which might lead to breakdown of the mucosal barrier). In the only previous adult case, ischaemic changes were seen but biopsy specimens did not show a vasculitis. Despite a thorough examination of the resection specimen in our case, we too found no evidence of vasculitis or of intravascular thrombosis.

Presentation of pneumatosis cystoides intestinalis with isolated pneumoperitoneum is uncommon; the condition is usually diagnosed after symptoms such as altered bowel habit, rectal bleeding, passage of mucus, abdominal pain/discomfort, urgency or excessive flatulence[2].

After the exclusion of a treatable cause or occult malignant disease, the treatment of pneumatosis is largely symptomatic. Oxygen therapy leads to a relative decrease in the partial pressure of non-oxygen gases in the blood which in turn diffuse out of the cysts leading to resorption. Antibiotics and elemental diets can give temporary relief[2]. If pneumoperitoneum occurs without peritonitis, it can be managed non-operatively.

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